J Ped Surg Case Reports 2 (2014) 462–464

Contents lists available at ScienceDirect

Journal of Pediatric Surgery CASE REPORTS

journal homepage: www.jpscasereports.com

In need of a patch UP: Recurrent congenital diaphragmatic hernia presenting with a large pleural effusion☆

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ARTICLE INFO

Article history:
Received 29 January 2014
Received in revised form
9 September 2014
Accepted 9 September 2014

Key words:
Congenital
Diaphragmatic hernia
Pleural effusion
Pediatric

ABSTRACT

We report a case of recurrent congenital diaphragmatic hernia (CDH) presenting with a large unilateral pleural effusion. A 12-year old boy who had a left sided CDH repaired in the neonatal period, presented with fever, lethargy, and non-productive cough. Chest radiograph demonstrated a loculated pleural effusion. Computed tomography scan revealed recurrent herniation of abdominal contents. To our knowledge, this is the first reported case of a recurrent congenital diaphragmatic hernia presenting with large pleural effusion.

Congenital diaphragmatic hernia (CDH) is estimated to occur with an incidence of 1 in 3600 live births [1], with the majority presenting in infancy. Diagnosis may be made antenatally by screening prenatal ultrasound, or postnatally by plain radiographs and clinical presentation. Physical signs and symptoms are variable and can include neonatal cardiorespiratory failure with need for extracorporeal membrane oxygenation (ECMO) in the neonatal period, as well as gastrointestinal obstruction, and failure to thrive in cases of recurrence or delayed diagnosis [2]. The incidence of re-herniation following CDH repair ranges from 7% to 46% [3,4] depending on the closure modality. Variable radiologic findings associated with trans-diaphragmatic herniation may be confused with other conditions such as pleural effusion or in some cases pneumonia [1,5]. As a consequence, both the initial diagnosis of CDH and that of re-herniation may be difficult.

1. Case report

A 12 year-old boy, who presented to a community hospital after 5 days of progressive non-bilious vomiting, anorexia, chest pain, lethargy, and mild non-productive cough. Notable past medical history included an antenatally diagnosed left sided congenital diaphragmatic hernia in addition to dextrocardia. His diaphragmatic defect had been repaired on the sixth day of life after three days of ECMO and involved placement of a Gore-Tex patch in-situ over a large left posterolateral defect.

On this presentation, initial chest radiograph (CXR) showed findings suggestive of a left sided parapneumonic effusion (Fig. 1). The patient was started on IV antibiotics and transferred the 900 km to the British Columbia Children's Hospital. Over the following 3 days, he showed minimal improvement, with deteriorating CXR findings, increased oxygen demand, and persistent spiking fevers. A pigtail catheter was inserted into the left anterior chest under radiologic guidance, with early return of 500 cc of serosanguinous fluid and further drainage of 1200 cc on the ward. Culture of the fluid demonstrated no bacterial growth and the patient showed mild improvement in his respiratory status following drainage of the effusion. A computed tomography scan of the chest was subsequently performed to assess whether the patient might benefit from a VATS procedure, and demonstrated evidence of visceral re-herniation through the previous diaphragmatic defect (Fig. 2). General surgery was consulted, and the patient was taken to the operating room for repair of his recurrent hernia.

Intraoperatively, recurrence of the hernia was noted where patch had previously been tethered to the posterior rib cage.

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http://dx.doi.org/10.1016/j.epsc.2014.09.008

Fig. 1. Initial CXR demonstrating significant left sided opacity and moderate midline shift.

Other significant findings included multiple adhesions, an incarcerated gastric fundus, and a small gastric perforation with surrounding necrosis. There was no evidence the pigtail catheter had injured any viscera. The abdominal contents were reduced, gastric perforation primarily repaired and a new Gore-Tex patch placed. The patient's recovery was uncomplicated, and he was discharged home on the seventh post-operative day. Follow-up imaging revealed complete resolution of the left sided effusion (Fig. 3). At 2 years after repair of his recurrent CDH, he is being followed by physicians in his community and remains recurrence free.

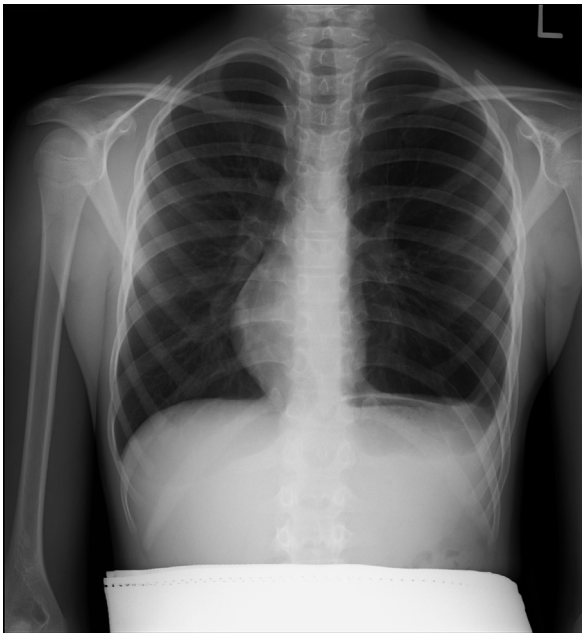

Fig. 3. CXR one year post-repair demonstrates complete resolution of effusion and no evidence of recurrence.

2. Discussion

While primary muscular closure is the preferred operative repair technique for CHD repair, it is not always appropriate in infants with large diaphragmatic defects. In up to 51% of patients with CDH, primary repair is not possible and requires a prosthetic repair [6,7]. Rate of recurrence with patch repairs has been reported between 41 and 46% [3,7] in comparison to 7–10% in those with primary closure [4]. Factors influencing likelihood of recurrence include patch material, fixation technique, intra-abdominal pressure and excess tension on the closure, usually related to size of the defect and available adjacent tissue and prosthetic patch size [8]. In addition to the increased risk for recurrence, slower early post-repair growth, and a higher incidence of gastro-esophageal reflux

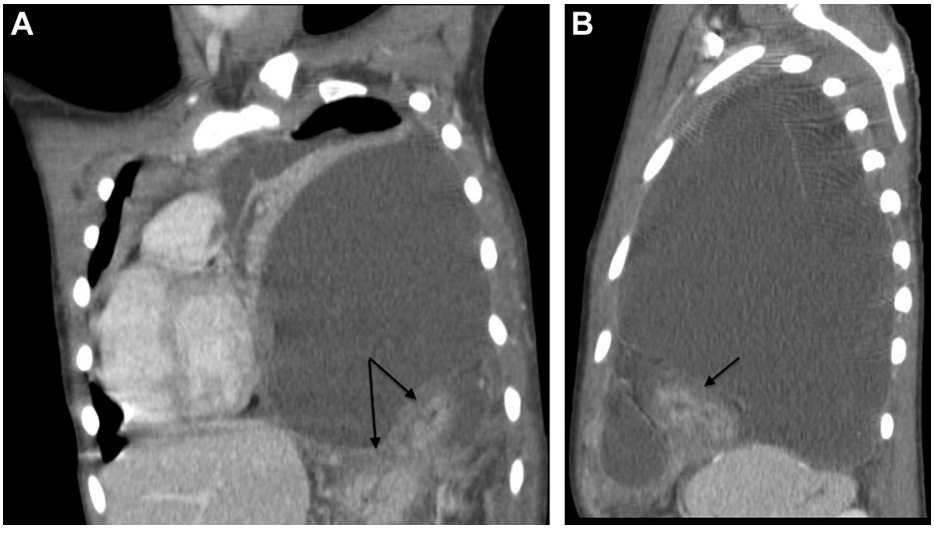

Fig. 2. CT scan of the chest showing large simple left thoracic fluid collection with evidence of trans-diaphragmatic herniation of abdominal contents. A) Arrows demonstrate stomach traversing the diaphragm; B) arrow demonstrates intra-thoracic gastric fundus.

have been reported in patients treated with patch compared to those undergoing primary repair [3,9].

Parapneumonic effusion as a presenting finding has been documented in traumatic diaphragmatic rupture in the adult population. There are also reported cases of clinically and radiologically misinterpreted late-presenting CDH as pneumonia with effusion; however, to our knowledge, there are no other reported cases of such a presentation in recurrent trans-diaphragmatic herniation from a congenital defect [1,10,11]. The case illustrates that recurrent diaphragmatic hernia should be considered in any patient with previous CDH repair presenting with parapneumonic effusion, as it has the potential to masquerade as a respiratory illness. This highlights the need for a complete and thorough pediatric history. Accompanying symptoms such as intestinal obstruction or other non-specific gastrointestinal complaints may support a diagnosis of recurrent herniation, but should not be relied on, so as not to delay diagnosis.

3. Conclusion

Early recognition of re-herniation is critical, as prolonged time to diagnosis may place the intrathoracic abdominal organs at risk for incarceration, necrosis and perforation. Ideally, regular surveillance will identify those who have recurred, before becoming symptomatic. Current recommendations are for serial postoperative radiographs in all patients who had their CDH repaired with a patch every 3 months in the fist year, one at 15–18 months post-repair and yearly thereafter [12]. It is possible that had our patient been followed in such a way, the recurrence may have been detected prior to symptoms [13]. Given that in patients with late presentations of CDH, misdiagnosis rates of up to 38.2% occur using chest radiography alone [11], further imaging modalities including ultrasound and computed tomography should be strongly considered in a patient with a history of repaired CDH and abnormal chest radiograph.

We have described an atypical presentation of recurrent congenital diaphragmatic hernia. This may present in a multitude of ways, and high index of suspicion must be maintained especially in patients with a history of patch repair and persistent respiratory or gastrointestinal symptoms. Additional care should also be taken to rule out recurrence in this population prior to invasive interventions to prevent inadvertent organ injury. Adherence to published guidelines for surveillance of CDH patients will allow identification and management of complications to mitigate against the development of further morbidity.

Conflict of interest statement

There are no financial or personal conflicts to disclose.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

References

[1] Wooldridge JL, Partrick DA, Bensard DD, Deterding RR. Diaphragmatic hernia simulating a left pleural effusion. Pediatrics 2003;112(6 Pt 1):e487.
[2] Elhalaby EA, Abo Sikeena MH. Delayed presentation of congenital diaphragmatic hernia. Pediatr Surg Int 2002;18(5–6):480–5.
[3] Jancelewicz T, Vu LT, Keller RL, Bratton B, Lee H, Farner D, et al. Long-term surgical outcomes in congenital diaphragmatic hernia: observations from a single institution. J Pediatr Surg 2010;45(1):155–60; discussion 160.
[4] St Peter SD, Valusek PA, Tsao K, Holcomb 3rd GW, Ostlie DJ, Snyder CL. Abdominal complications related to type of repair for congenital diaphragmatic hernia. J Surg Res 2007;140(2):234–6.
[5] Berman L, Stringer D, Ein SH, Shandling B. The late-presenting pediatric Bochdalek hernia: a 20-year review. J Pediatr Surg 1988;23(8):735–9.
[6] Clark RH, Hardin Jr WD, Hirschl RB, Jaksic T, Lally KP, Langham MR, et al. Current surgical management of congenital diaphragmatic hernia: a report from the Congenital Diaphragmatic Hernia Study Group. J Pediatr Surg 1998;33(7):1004–9.
[7] Moss RL, Chen CM, Harrison MR. Prosthetic patch durability in congenital diaphragmatic hernia: a long-term follow-up study. J Pediatr Surg 2001;36(1):152–4.
[8] Hajer GF, vd Staak FH, de Haan AF, Festen C. Recurrent congenital diaphragmatic hernia; which factors are involved? Eur J Pediatr Surg 1998;8(6):329–33.
[9] Valfre L, Braguglia A, Conforti A, Morini F, Trucchi A, Iacobelli BD, et al. Long term follow-up in high-risk congenital diaphragmatic hernia survivors: patching the diaphragm affects the outcome. J Pediatr Surg 2011;46(1):52–6.
[10] Booker PD, Meerstadt PW, Bush GH. Congenital diaphragmatic hernia in the older child. Arch Dis Child 1981;56(4):253–7.
[11] Baglaj M, Dorobisz U. Late-presenting congenital diaphragmatic hernia in children: a literature review. Pediatr Radiol 2005;35(5):478–88.
[12] Section on Surgery and the Committee on Fetus and Newborn, Postdischarge follow-up of infants with congenital diaphragmatic hernia. Pediatrics 2008;121(3):627–32.
[13] Kadian YS, Yattan KN, Verma M, Kajal P. Congenital diaphragmatic hernia: misdiagnosis in adolescence. J Indian Assoc Pediatr Surg 2009;14(1):31–3.